# Visually guided step descent in children with Williams syndrome

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**Abstract**

Individuals with Williams syndrome (WS) have impairments in visuospatial tasks and in manual visuomotor control, consistent with parietal and cerebellar abnormalities. Here we examined whether individuals with WS also have difficulties in visually controlling whole-body movements. We investigated visual control of stepping down at a change of level in children with WS (5-16 year olds), who descended a single step while their movement was kinematically recorded. On each trial step height was set unpredictably, so that visual information was necessary to perceive the step depth and position the legs appropriately before landing. Kinematic measures established that children with WS did not use visual information to slow the leg at an appropriate point during the step. This pattern contrasts with that observed in typically developing 3- and 4-year old children, implying severe impairment in whole-body visuomotor control in WS. For children with WS, performance was not significantly predicted by low-level visual or balance problems, but improved significantly with verbal age. The results suggest some plasticity and development in WS whole-body control. These data clearly show that visuospatial and visuomotor deficits in WS extend to the locomotor domain. Taken together with evidence for parietal and cerebellar abnormalities in WS, these results also provide new evidence for the role of these circuits in the visual control of whole-body movement.

Williams syndrome (WS) is a genetic disorder estimated to affect about 1 in 7,500 of the population (Stromme, Bjornstad & Ramstad, 2002). It is associated with genetic deletion on chromosome 7, band 7q11.2. This typically centres on the elastin gene (ELN) and includes a surrounding region of genes, pseudogenes and markers (Bellugi, Lichtenberger, Mills, Galaburda, & Korenberg, 1999). While ELN deletion is associated with the heart problems found in WS, the surrounding genes seem to be of primary importance for the other characteristic physical and cognitive features of the syndrome. The functional relationship between these genes and specific cognitive deficits can be studied either by correlating functional abilities with microdeletions in individual patients; or by correlating genetic expression with cognition in the typical WS population. Recent results suggest that the gene STX1A may particularly associated with learning and memory deficits in WS (Gao *et al*, 2010), but we are only beginning to understand the exact genetic bases of the WS phenotype.

Individuals with WS typically have an uneven cognitive profile, in which communicative functions are relatively spared but visuospatial abilities are impaired (Atkinson, Anker, Braddick, Nokes & Mason, 2001; Bellugi *et al*, 1999; Frangiskakis *et al*, 1996; Mervis, Robinson & Pani, 2000). An important question is whether these visuospatial impairments extend to whole-body visuomotor control, including walking (Atkinson *et al*, 2001; Withers, 1996). Behavioural and neural data suggest that this may be the case, but it has not been directly tested. It is an area of clear practical importance to individuals with WS. Furthermore, since the neural bases of visuomotor control in walking are poorly understood in humans, studying individuals with WS may provide a

valuable contribution to understanding the foundations of this system and its development.

*Visual control of manual action in WS*

Existing data indicate that individuals with WS may have atypical functioning in the dorsal stream (Atkinson *et al*, 1997), which controls action plans on the basis of visual information (Milner & Goodale, 1995). For WS adults and children motion processing, subserved by the dorsal stream, is typically worse than form processing, subserved by the ventral stream (Atkinson *et al*, 1997; 2006). Importantly for the current study, children and adults with WS have deficits in a range of manual tasks that require visual information to guide and calibrate actions. Children with WS were more impaired when posting a card through an oriented slot than when matching the card with the slot's orientation (Atkinson *et al*, 1997). Impairments relative to controls were also shown on a visuomotor grasp-scaling task (Newman, 2001). Both these findings suggest that children with WS have difficulty in controlling movement (hand size or orientation) on the basis of visual features. Similarly distance scaling has been examined in manual movements of adult participants with WS (Elliott, Welsh, Lyons, Hansen & Wu, 2006). Participants with WS made slower movements and more online corrections than typically developing adult participants, participants with Down syndrome, or participants with delays of unknown aetiology. Furthermore, peak movement velocity did not scale to target distance as it did for participants in the other groups. Again this lack of adaptation suggests a profound difficulty in integrating visual information into action plans in WS: however, these studies are specific to the manual domain (Hocking, Rinehart, McGinley, Moss & Bradshaw, 2011).

As well as this behavioural data, there is evidence to suggest that dorsal/parietal neural structures are atypical in individuals with WS. Neuroimaging shows abnormal white matter in the parietal lobes of infants with WS (Mercuri *et al*, 1997); children with WS show abnormally low grey matter concentration in the parieto-occipital lobe (Boddaert *et al*, 2006); and young adults show decreased parietal lobe volume (Eckert *et al*, 2005; Reiss *et al*, 2004). These structural abnormalities can be directly linked to functional deficits. A group of adults with WS who had reduced gray matter volume in the parieto-occipital/ intraparietal sulcus region also showed lower activation than controls in a dorsal stream region just upstream of the structurally damaged area during performance on visuospatial and visuomotor tasks (Meyer-Lindenberg *et al*, 2004). Together these behavioural and neural data suggest a fundamental difficulty in controlling manual action on the basis of visual information, which results from parietal difficulties. The present study investigates whether this difficulty extends into the whole-body, locomotor domain.

*Whole-body control in WS*

Basic locomotor problems are consistently reported in WS (e.g. Atkinson *et al*, 2001; Chapman, du Plessis & Pober, 1996; Elliott *et al*, 2006). Adults with WS walk more slowly than healthy controls, primarily due to decreases in stride length rather than cadence (Hocking, Rinehart, McGinley & Bradshaw, 2009). This study also found an increased variability in base of support and stride length in WS participants. These results do not directly reflect on the visual control of walking, but a brief report by Withers (1996) documents several problems in this group including reluctance to cross from one textured surface to a new one and problems walking down stairs. These situations both

involve the incorporation of visual information about the environment into the locomotor plan. More recently, Hocking, McGinley, Moss, Bradshaw & Rinehart (2010) found that adults with WS show an unexpected pattern of walking when visual cues are provided. Such cues can increase walking speed, for example in Parkinson's disease (Azulay *et al*, 1999). However in WS they cause reductions in stepping frequency and speed. This indicates that visuomotor difficulties may extend into whole-body control in adults with WS. However more study of this is warranted, particularly in children with WS.

Studying WS may also increase our understanding of visually guided whole-body movements, the neural basis of which is poorly understood and derived mostly from animal data (Georgopoulos & Grillner, 1989; Hashimoto, 2006). A working hypothesis is that visually guided whole-body control depends on both parietal and cerebellar structures: evidence from the cat suggests that parietal circuits may allow a modulation of rhythmic locomotor patterns on the basis of visual inputs (Drew, Andujar, Lajoie & Yakovenko, 2008); and both human and animal data suggest that cerebellar circuits are key for balance and coordination (Morton & Bastian, 2007). Studying WS may allow a link to be made between this hypothesis of neural function and observable behaviour in human subjects. We have already reviewed the literature suggesting that dorsal/parietal circuits are atypical in WS. Recent work also confirms cerebellar abnormalities in WS. Cerebellar signs including oculomotor difficulties (van der Geest *et al*, 2004) and awkward gait (Pober, 2010) are reported; individuals with WS also have poorly regulated, highly variable stride times (Hocking *et al*, 2010). Regions of the cerebellum are enlarged in children (Jones *et al*, 2002; Mercuri *et al*, 1997; Pober & Foliano, 1995) and adults with WS (Schmitt, Eliez, Warsofsky, Bellugi & Reiss, 2001). Other relevant

structures such as the basal ganglia may also be impaired during development in WS (see Hocking, Bradshaw & Rinehart, 2008). Showing a correlation between known parietal /cerebellar damage and functional impairments in WS would therefore add support to the hypothesis that these circuits contribute to whole-body control in humans.

*Visual control of step descent in WS*

The present study uses a step descent paradigm to investigate visually guided locomotion in children with WS. The descent of steps and kerbs is commonly reported as a problem by WS families. Stepping down from a surface of one height to another is a crucial component of everyday locomotion; however, it is a complex movement. Detailed kinematic and kinetic analyses have shown that it involves characteristic patterns of joint flexion and extension (McFadyen & Winter, 1988; Protopapadaki, Drechsler, Cramp, Coutts & Scott, 2007; Riener, Rabufetti & Frigo, 2002), which culminate in the extension of the leg joints in preparation for landing (McFadyen & Winter 1988; Zachazewski, Riley & Krebs, 1993). EMG also shows characteristic bursts before landing (Craik, Cozzens & Freedman, 1982; Santello, 2005). One important component of step descent is visuomotor control. A significant seam of research has studied how movements of the lower limbs, or whole-body, can be adapted to the dimensions of the environment (McCarville & Westwood, 2006; Patla & Goodale, 1996; Reynolds & Day, 2005). In step descent, certain parameters of the movement must be set according to the height of the step one is descending (Buckley, MacLellan, Tucker, Scally & Bennett, 2008; Cowie, Braddick & Atkinson, 2008; Cowie, Atkinson & Braddick, 2010). Using visual information to set these parameters correctly is a crucial part of making a safe step down and if visual input is blurred or the eyes covered then the adaptation of these parameters

to step height is diminished (Cowie *et al* 2008; Timmis, Johnson, Elliott & Buckley, 2009).

In the current paradigm participants descend a single step whose height is unpredictably shallow, medium, or deep. Visual control is demonstrated when participants use visual information about the step's depth to scale the values of particular kinematic variables during descent. We have shown that, with normal vision available, adults and typically developing three- and four-year-olds demonstrate visual control of stepping-down in this paradigm (Cowie *et al*, 2008, 2010). Using identical equipment and stepping procedures to those employed in Cowie *et al* (2010), the present study examined how well children with WS scaled movement parameters to step depth ('riser height'), and thus how well they were able to incorporate visual information into the whole-body motor plan during stepping-down.

We also investigated whether simple impairments in low-level motor control or visual functioning would predict stepping performance in WS. Perhaps the most relevant motor skill for step descent is dynamic balance, which depends on muscle strength as well as the use of sensory inputs, and provides a base from which to carry out controlled movements in a step descent task. Balance was assessed using a subtest from a standardised motor assessment battery, the Movement ABC (Henderson & Sugden, 1992). Low-level visual problems are frequent in WS: as many as half these individuals show refractive errors (usually hyperopia); and fail standard tests for stereo vision (Atkinson *et al*, 2001; van der Geest *et al*, 2005). To assess low-level visual functions we measured both acuity and stereo ability, though a previous study found no correlation

between parent-reported problems with stairs and stereo vision performance in children with WS (Atkinson *et al*, 2001).

We hypothesised that children with WS would be unable to scale movements to step height, and that movement would significantly contrast with behaviour of typically developing children. We further hypothesised that this would not be fully explained by deficits in basic balance, visual acuity or stereo vision, but would rather reflect deficits in higher level visuomotor processing.

## *Method*

### Participants

Sixteen children with Williams syndrome (5 - 16 years, mean 9.58, s.d. 2.9 years, ten males) took part in the study. They were recruited from a national database. 14/16 were positive on the fluorescent in situ hybridization (FISH) test, which confirms a deletion of the elastin gene on chromosome 7. Two children (the youngest and the oldest in the sample) had not taken the test, but had a clear WS phenotype and had been diagnosed with WS by medical practitioners. Prior to testing all participants (or their caregivers, as appropriate) gave informed consent. The protocol was approved by the local ethics committee and carried out according to the principles laid down in the 1964 Declaration of Helsinki.

### Equipment

A simple 'step' was constructed by vertically stacking 18mm planks of wood into two piles, constituting an 'upper' and a 'lower' platform. The height of the step's upper platform, which participants started on, was constant across trials. This meant that the

step up to the upper platform on each trial was no guide to the depth to be descended. Rather, riser height was varied by changing the height of the lower platform, which participants stepped down to. We recorded kinematic data using a 6-camera motion tracking system (SMART, Milan) operating at 60 Hz. Cameras surrounding a 1.6m x 1.6m x 1.6m testing area allowed accurate 3-D reconstruction of marker positions. Reflective markers were placed on the corners of the upper and lower steps, and on 8 lower-body locations. On each leg these were the lateral epicondyl of knee, ankle (lateral malleolus), toe ($2^{nd}$ metatarsal head), and heel (most posterior point of calcaneus, at the height of the $2^{nd}$ metatarsal head). Participants were barefoot and wore shorts to allow easy camera viewing of the kinematic markers.

**Procedure**

The task was to take one step down from the upper to the lower platform, with the lead and then the trailing leg, ending with feet side-by-side. A game was introduced between trials to motivate participants (Cowie *et al*, 2010). On completing a step down children were rewarded by choosing a picture of an animal to stick on a 'jungle' wall. Participants first practised walking with markers fitted and had one or two practice trials stepping down a step of random height. Because children with WS can become very anxious in 'testing' situations, parents were asked to judge whether the child seemed to be stepping as they did normally, and the experiment started when parents and experimenter judged the child to be comfortable with the markers and the task. Children walked to the upper platform with eyes closed, accompanied by the experimenter. When on the upper platform, they heard two 'boing' sounds, separated by 2 seconds. On the first sound they opened their eyes and looked at the step. On the second sound they stepped down.

## Design

Riser height (8, 16, 24% leg length) was varied within-subjects. Leg length was measured as the distance from greater trochanter to lateral epicondyl to lateral malleolus. Each participant completed 15 trials, in 5 blocks. Each block contained one trial at each of the 3 riser heights in pseudorandom order for each participant.

## Additional tests and questionnaire

As well as completing the stepping-down task, participants were tested on a battery of standard tests to assess different aspects of visual, cognitive and motor function. The description and scoring of these are shown in Table 1. Parents also answered a series of questions about their child's walking ability, with particular regard to stair descent. This short questionnaire was developed by the authors to explore the relationship between parental reports of home stepping, and stepping down ability as measured in the laboratory. Questions and scoring are shown in Appendix A.

## Movement kinematics and analysis

Data from the leading leg were analysed. We exported the 3D position coordinates of each marker in order to calculate the dependent measures. The raw 3D position data were first filtered with a 2$^{nd}$ order low-pass Butterworth filter, cut-off frequency 10Hz. We extracted several measures from each trial: kneedrop, toedrop, toeheight (speedpeak), maximum toespeed, and mean toespeed. These are described in detail in Cowie *et al* (2010) and illustrated here in Figure 1. During a step down, the calf swings outwards to a peak. It must then swing back in again: for a safe, efficient landing this must be done earlier for a shallower step. For this reason our first two measures (kneedrop and toedrop) focus on the control of this swinging-in process and its relation to step height. We first

considered the positional difference between knee and ankle positions in the forward dimension. We defined the calf's peak outward swing as the point at which that difference reached a minimum or plateau. Measures were then defined as follows. Kneedrop is the distance descended by the knee during the leg's outward swing. Toedrop is the distance descended by the toe from its starting position on the upper platform to its position at peak swing. Our other measures focus on the control of toe speed during the step down. Toeheight (speedpeak) is the vertical distance the toe rises from its starting position on the upper platform to the point where it reaches its maximum speed. Mean toespeed and maximum toespeed are the mean and maximum values of the toe's speed during the step down. After analysis, we averaged measures from the 5 trials at each riser height for each individual participant. Our previous work shows that in healthy adults all five kinematic measures scale to step height; in typically developing children with full vision of the step, kneedrop, toedrop and toeheight (speedpeak) measures scale to step height (Cowie *et al*, 2010).

Our analyses addressed several questions. First, do individuals with WS scale movement parameters to step height? To answer this question we conducted a one-way ANOVA on the mean of each measure with main effect 'riser height'. Our second analysis asks how the development of stair descent in individuals with WS compares to typically developing children. The group of TD children combined all the 3 and 4 year olds who yielded complete datasets in Cowie *et al*, 2010. This comprised nine 3 year olds (mean age 3.48, s.d. 0.06 years, 4 males) and nine 4 year olds (mean age 4.51, s.d. 0.05 years, 5 males).

Because of the uneven cognitive profile in WS, it is not possible to assign an overall mental age equivalent for this group. Setting a match based on spatial tasks risks underestimating mental age, while setting a match based on verbal tasks risks overestimating (Jarrold & Brock, 2004). Rather than using a matched group design, we have chosen the TD comparison group based on our knowledge of the development of movement scaling ability in the typical population. In our previous work we found no significant development in movement scaling under conditions of normal vision between 3 years and adulthood. Whilst this suggests that a TD comparison group of 3-year-olds or of adults would yield the same pattern of results, comparison of a TD adult group to the performance of WS children would make it difficult to rule out group differences in experience and in mental age as contributors to any group differences. The most conservative method, therefore, is to choose a TD comparison group at the lowest end of the developmental trajectory that we have been able to measure using the current task. As such, the TD comparison group consists of 3-year-olds and 4-year-olds. Although our previous data indicate that children of this age have reached the endpoint of the developmental trajectory in their ability to scale movements to step height, they have a lower chronological age than the WS children (and one assumes, reduced experience of stair descent), and have a mental age which is at or below that of the WS children on both verbal and performance measures (WS mean BPVS mental age: 5.8 years; WS mean WPPSI Object assembly mental age: 4.5 years). Thus any impairment in WS performance on the experimental task cannot be attributed to group differences in chronological age or mental age. All but one child in the WS group had BPVS mental age above 3 years: this child was excluded from these analyses. We conducted repeated

measures ANOVAs on kinematic parameter means with the factors riser height (8, 16, 24% leg length) and group (WS, TD children). A second set of ANOVAs determined whether within-person variability was different between WS and TD groups.

Our third question for analysis was whether scaling performance within the WS group related to other developmental measures. To answer this question we conducted correlation analyses to assess how scaling performance depended on a number of potentially relevant factors for the WS group. Pearson correlation was used for interval predictors and Spearman's rank correlation for ordinal predictors. For all analyses the dependent variable was the slope of the best-fit riser height *vs* toedrop function, calculated for each individual. This kinematic variable has proved to be a sensitive marker of visuomotor control in step descent in previous developmental experiments, indicating a fine level of control which seems difficult for younger children to achieve, for example in altered visual conditions (Cowie *et al*, 2010).

Finally, we asked if performance on our stair descent task correlated with parent-reported problems with stairs, and with other locomotor problems. To answer this question we assessed how toedrop scaling performance (the slope of the line relating step height to toedrop) related to questionnaire answers. Spearman's rank correlation was used for ordinal variables, and independent samples t-tests (equivalent to point-biserial correlation) for dichotomous variables. Questionnaire items 6 (difficulties with step ascent), 7 (difficulties with descent) and 10 (backward stepping) did not have a large enough spread of values to analyse. The different variables are of course likely to be intercorrelated, but we did not have sufficient data in our sample to perform multivariate analysis.

## *Results*

### Questionnaire data

Eleven participants yielded questionnaire data. All these children regularly used a staircase at home. All but one at some time had had significant amounts of physiotherapy or undertaken special balance or walking exercises; one had had a small amount of physiotherapy. Thus the locomotor background of our sample was quite uniform.

8/11 children were reported to be hesitant at kerbs; 2 were not and one had been as a younger child. 3/11 were hesitant at a change from one texture to another; 7 were not and one had been as a younger child. This tallies with the figure of 30% reported in Withers (1996). 5/11 were hesitant while walking on sand. These problems have often been reported anecdotally and this small-scale survey suggests that their visuomotor basis is worth investigating more thoroughly than has so far been done. The vast majority of the group had specific problems with stairs: 10/11 families reported that the child had problems with descent and 5/11 with ascent. Specifically, 1/11 always went down stairs backwards; 1/11 sometimes; 2/11 never and 7/11 had done so in the past. The rail was always used by 6/11; often by 3/11 and sometimes by 2/11. Catch-up stepping was always used by 6/11, often by 4/11 and never by 1/11.

### Kinematic data

Example traces of lower limb movement are shown in Figure 2. From these we extracted the key kinematic parameters necessary to address our hypotheses, as follows.

***Do individuals with WS scale movement parameters to step height?***

Group mean values of each kinematic variable are presented in Figure 3. Repeated measures ANOVAs showed significant linear effects of riser height on the group mean values of kneedrop ($F[1,15] = 22.5$, $p < .001$), toedrop ($F[1,15] = 25.0$, $p < .001$), and mean toespeed ($F[1,15] = 12.5$, $p < .004$). These effects of riser height show that the WS group scale these variables to step height on the basis of visual information. However there were no significant effects of riser height on toeheight (speedpeak) ($F[1,34] = 0.14$, $p > .7$) or maximum toespeed ($F[1,15] = 3.9$, $p < .07$). These variables are therefore not scaled to step height in the WS group.

***Is development of stair descent in individuals with WS delayed relative to typically developing children?***

Table 2 shows that for kneedrop and toedrop there were significant effects of riser height but no significant effects of group or interactions. Toeheight (speedpeak) was also significantly scaled to riser height, but a riser height x group interaction shows that the extent of scaling was different for the WS and TD groups. Therefore, the WS group did not scale toeheight (speedpeak) in the same way as typically developing children (Figure 3). Likewise mean toe speed was significantly scaled to riser height, but with an effect of group such that children with WS stepped down more slowly than the TD group (see Figure 3). Finally, maximum toe speed was not significantly affected by riser height or group. Within-person variability is shown in Figure 4. The WS group had significantly less within-person variability for mean toespeed (Fig 4, Table 3). Variability in toedrop and kneedrop increased with step height. There were no group differences in within-person variability for other parameters.

In summary, children with WS scaled some of the same movement parameters as TD children. For example kneedrop and toedrop measures were scaled. However, the profile of the WS group was atypical. For toeheight (speedpeak), children with WS did not scale movement to riser height in the same manner as TD children, but rather reached peak velocity at a constant position irrespective of the step they were descending. Additionally children with WS had a lower, less variable mean foot speed than TD 3 and 4 year olds.

***Do other developmental measures predict performance in children with WS?***

Figure 5 plots the slope of the best-fit riser height *vs* toedrop slope, calculated for each individual, against each of seven variables. Because of time restrictions and testing demands not all children completed all tests. For each test, data are shown for the full dataset obtained. BPVS equivalent age significantly correlated with scaling performance ($r$ = .559, n=15, p = .03). However scaling performance was not significantly correlated with chronological age ($r$ = .446, n=15, p = .096), dynamic balance ($\rho$ = .485, n=10, p = .156), or age equivalence on the Object Assembly test ($r$ = .114, n=13, p = .711). Finally the visual factors acuity and stereo acuity were not significantly correlated with scaling performance (Binocular acuity: $\rho$ = -.487, n= 7, p = .268; Monocular Acuity in worst eye: $\rho$ = .175, n= 13, p = .566; Stereo Acuity: $\rho$ = -.292, n= 15, p = .291).

***Do parent-reported locomotor problems predict performance in children with WS?***

Questionnaire reports of strategies on stairs (rail use; catch-up stepping) did not significantly correlate with scaling performance (Rail use: $\rho$ = .079, n= 10, p = .828; Catch-up: $\rho$ = .125, n= 10, p = .731); neither did hesitancy at kerbs (t(8) = -1.34, p = .258) or on sand (t(8) = -.28, p = .785). Interestingly, reported hesitancy at texture

changes was significantly correlated with the ability to scale movements to step height ($t(8) = -2.895$, $p = .02$).

## Discussion

In this study, we examined the extent to which children with WS scaled their stepping movements to the height of step they were descending. Based on previous findings we hypothesised that children might be unable to scale movements to step height, and that this would contrast with typically developing children. We further hypothesised that this would not be fully explained by balance or acuity problems, but rather by high-level visuomotor difficulties. These predictions were supported by the study.

*Movement scaling in WS and TD groups*

We found that children with WS showed some degree of movement scaling to step height for several of the kinematic parameters scaled by adults (Cowie *et al*, 2008) and TD children (Cowie *et al*, 2010). This suggests that for children with WS, the broad purpose and pattern of movement in this task is the same as for TD children and adults. However, children with WS did not scale toeheight (speedpeak) to step height. Rather, their movement slowed down at the same point irrespective of step height. This atypical pattern confirms our first hypothesis of a significant lack of scaling in children with WS. As we discuss later with reference to the TD group, this most likely reflects a specific difficulty in incorporating visual information into the action plan.

Our second hypothesis was that children with WS would produce different movements to TD participants. We found that stepping movements in the WS group were slower than in the TD group. In addition, TD children of three years scale toeheight (speedpeak) to riser height: in other words they set the position at which they reach peak

speed relative to the step. In contrast, as discussed above this is not true of children with WS, who begin slowing down at the same point irrespective of step height. This significant difference between the two groups confirms that, for our sample, children with WS scaled movements differently to TD participants.

As explained in the methods section, we assumed that our group of WS children should have been performing at least as well as the 3-4-year-old TD group. One WS participant had a verbal mental age of less than three years and was removed from all statistical analyses of kinematic measures. The remaining WS group had an average verbal mental age (BPVS equivalent) of 6.0 years, higher than the 3-4-year-old TD group. In this respect the TD group represents a lower bound of development with which a WS group might be compared. However, since verbal performance is typically higher than spatial performance in WS groups (Bellugi *et al*, 1999), this comparison does risk disadvantaging the WS group. Considering instead spatial performance, two WS participants did not complete the WPPSI Object Assembly test, but the mean mental age equivalence for the remainder of the sample was 4.5 years. Only one child's equivalent age was below 3 years. Therefore the mental age of the WS sample was within or above the range of the TD group on spatial as well as verbal measures. We can therefore conclude that the significant difference between WS and TD groups results from meaningful comparisons, and highlights important differences in whole-body visuomotor control over and above the known pattern of impairments in WS.

*Explaining atypical performance in the WS group*

We examined a number of predictors for performance in the WS group. Some of these, in particular the unvalidated questionnaire items and the test items with reduced datasets,

must be considered preliminary. However we systematically address a number of possible predictors here and argue for a deficit in whole-body visuomotor control in children with WS.

Variation within the WS group was not well explained by impairments of acuity, stereo acuity, or dynamic balance. This is consistent with work on WS reaching, where detailed measures of visual ability do not predict the scaling of reaches to target distance (Newman, 2001; van der Geest *et al*, 2005). This highlights the separability of perceptual and visuomotor processing, consistent with the hypothesis of distinct visual pathways subserving perception and action and its extension to stepping paradigms (see Patla & Goodale, 1996). Furthermore, the movement pattern of the WS group is not suggestive of muscle weakness, though this is an issue which could be assessed more directly in future studies. We are therefore able to rule out some lower-level factors as predictors of performance in the WS group. Two remaining hypotheses might account for the movement patterns we observed. We will term these the 'visuomotor deficit' hypothesis and the 'caution' hypothesis.

The visuomotor deficit hypothesis is that the atypical stepping we observed was caused by a deficit of visuomotor processing. There were two noticeable patterns in WS stepping. First the WS group started decelerating at the same point irrespective of step height. In other words they did not use visual information about step height to optimise the movement pattern captured by the variable 'toeheight (speedpeak)'. Second, toedrop scaling depended on verbal mental age (Figure 5). For children with lower mental age, the leg began swinging in to land early in the step irrespective of visual information about riser height. Both these results suggest that the WS group have a core impairment in

using visual information about step height to control their stepping movements. This 'visuomotor deficit' hypothesis is consistent with what we know of parietal neuronal changes in WS (Meyer-Lindenberg *et al*, 2004; Reiss *et al* 2004), as well as WS performance in manual visuomotor tasks (Hocking *et al*, 2011) and the limited evidence we have on whole-body control in WS (Hocking *et al*, 2010). Though the neural basis of visually controlled locomotion is uncertain (Cowie *et al*, 2010), it is likely to have strong contributions from the same dorsal/parietal network that controls visually guided reaching and grasping (Hashimoto, 2006). As reviewed in the introduction, there is extensive evidence that these areas have atypical structure and / or function in WS (see Hocking *et al*, 2008). Behavioural studies also show that upper limb reaching movements in WS are not adapted to target distance. Reaches have long periods of deceleration (Elliott *et al*, 2006; Newman, 2001; van der Geest *et al*, 2005), in the same way that the leg begins swinging in early in our stepping down task. Furthermore, van der Geest *et al* found better reach scaling in the small subgroup of their patients who did not have self-reported difficulties on stairs. Our results are therefore highly consistent with the existing upper limb literature, and suggestive of a core visuomotor deficit in WS which extends across the reaching and locomotor domains.

A second hypothesis to consider in explaining WS performance is that WS participants were overly cautious in their stepping down. According to this hypothesis, the core impairment in WS step descent is a fear of stairs, causing a hesitant stepping pattern. Certainly we found cautious behaviour in the WS group. Questionnaire items revealed a high degree of caution in natural situations. Kinematic analyses show low mean speeds and variance which suggests a carefully controlled, safe stepping pattern.

Likewise WS children of lower mental age began swinging the leg in to land quite early, irrespective of step height (reflected in the toedrop variable, see Fig 3). These results are consistent with WS participants descending stairs more cautiously than controls. However the major question of interest was whether children with WS scaled their movements appropriately for a visually specified step height. In this case the difference between WS and TD groups cannot be accounted for simply by the hypothesis that WS participants pursued a more cautious strategy. The toeheight (speedpeak) variable, whose scaling was significantly different between WS and TD groups, did not follow a cautious pattern. Cautiousness would principally involve the avoidance of high-speed collisions with the step. To do this, one should start slowing down early in the step, so that if the foot contacts it unexpectedly early there is a minimal chance of hitting it with a high speed. If participants with WS had been following this strategy, then toeheight (speedpeak) should have been on average lower in the WS group than the TD group. However this was not the case – there was no main effect of group on this variable, indicating that on average WS participants began slowing down at a medium or relatively late point irrespective of step height (see Fig 3). This is most indicative of a purely visuomotor problem where visual information about riser height was simply not integrated with motor plans to optimise stepping.

If we accept a visuomotor deficit hypothesis to explain performance, what might have caused such a deficit in the WS group? Evidence reviewed in the introduction to this paper suggests that several regions of the brain important for action planning and control develop atypically in WS. The dorsal stream, parietal cortex and cerebellum are particularly implicated. However, little is known about whether these areas control

whole-body movements as well as manual behaviour. Importantly the present results show a deficit in visuomotor whole-body control in WS, a group with known parietal damage. Together with the work from other sources (Hocking *et al*, 2010; Hanakawa, Fukuyama, Katsumi, Honda & Shibasaki, 1999) this points to dorsal-parietal networks as the key components for action control, both whole-body and manual. As new technologies permit measurement of children's behaviour in increasingly naturalistic situations (Pereira, James, Jones & Smith, 2010; Franchak & Adolph, 2010), so understanding whole-body control and its basis is an important new direction for developmental research as a whole.

Future studies should investigate the exact nature of the visuomotor deficit in WS. For example, scaling problems may have occurred at the planning stage of the movement or during its online control. In a pointing task, van der Geest *et al* (2005) found that participants with WS were more impaired than controls when visual feedback was removed, overshooting a manual target they were aiming for. This implies that online control may be more important for this group than for TD children, and that planning may be impaired. However, Elliott *et al* (2006) included an open-loop condition which yielded equivocal results. During step descent young children are more dependent than older children on online vision (Cowie *et al*, 2010). In some ways WS performance resembles the performance of TD children with feedback removed. Since these parameters were the least well scaled in the WS group it may be that a key deficit in this group is in using visual feedback to control movement parameters when necessary. It would be interesting for future studies to examine performance in the WS group with feedback removed.

Our analysis has concentrated on certain aspects of the stepping movement which allow us to specifically address the hypotheses set out in the introduction. Future work may address different aspects of the stepping movement, the complexity of which is evident from Fig 2. For example examining the fixed points and error patterns of a movement can shed light on the reference frames that participants are using to control it (Burnod *et al* 1999). One observation from our data is that the knee rises to its peak rather sharply in WS participants in comparison with the TD group. This may suggest that knee is being used as a fixed point in WS. This kind of strategy has been found in other neurodevelopmental disorders, for example in Angelman syndrome (Dan, Bouillot, Bengoetxea, Boyd & Cheron, 2001). However, recent work has demonstrated the complexity of determining the references used for movement, since a movement may be carried out with multiple reference frames (McGuire & Sabes, 2009) and determining these is not within the scope of this paper.

Head movement may be particularly interesting to look at in future studies. A number of brainstem reflexes (eg opto-cervical, vestibulo-cervical) stabilise the head during whole-body movement, allowing for a head-centred visual reference frame. In typically developing infants there is an increasing tendency towards stabilisation in the first 6 months of life (Brown, Omar & O'Regan, 1997). In several developmental disorders, however, head movement is not stabilised. For example in spastic diplegia and Angelman Syndrome the head flexes when squatting and extends when straightening up (Dan *et al*, 2001). In these disorders, the lack of a stable visual reference frame may play a role in deficits of visual perception (Dan *et al*, 2000). Likewise in WS it is possible that the visuomotor difficulties we describe may result in part from an unstable visual frame

of reference caused due to a moving head. Recent work has shown that gaze is directed down at a staircase during descent (Miyasike-daSilva, Allard & McIlroy, 2011; Zietz & Hollands, 2009); but there has been little analysis of head movement during step descent and indeed further work is needed to investigate this control in both typically and atypically developing groups.

*Age trends and practical implications*

There was no significant increase in performance with chronological age, but the data do suggest some developmental change in the ability to scale movement parameters during step descent across the age range studied (5 – 16 years). The current findings suggest that the extent of scaling performance in step descent is predicted by verbal mental age. This suggests a higher level factor contributing to stair descent ability. In a similar manner, Hocking *et al* (2009) found that lower Performance IQ was associated with reduced stride length in adults with WS during level ground walking. There is an apparent discrepancy between this study and our results in whether gait performance is better predicted by verbal or performance IQ. However, the studies employed different tasks (level walking *vs* step descent), and the reduction in stride length captures a more basic locomotor deficit than the scaling ability we are measuring here. Further longitudinal studies using a broader age range that incorporates both children and adults with WS will be necessary to further explore whether visually-guided locomotion may undergo important developmental changes throughout the lifespan (Karmiloff-Smith, 1998). The finding also suggests plasticity in the developing WS brain, where after early anomalous development in parietal or cerebellar areas, additional regions could increasingly come to play a role in visually guided stepping. Future research should model the balance between

anomalous neural development and neural plasticity in WS. Since all the children we assessed had undergone some form of physical therapy we cannot determine the effects of this in the present sample. Nevertheless, the current findings provide an impetus for future work which develops appropriate physical therapies for individuals with WS.

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**Figure &table legends**

**Figure 1:** Kinematic parameters. Reproduced from Cowie *et al* 2010.
**Figure 2:** Sagittal plane knee (upper) and toe (lower) movement, during descent of a high step. First trial from each participant shown.
**Figure 3:** Mean movement parameters in the WS and TD groups (TD data from Cowie *et al*, 2010). Group means and standard errors shown. LL: leg length.
**Figure 4:** Within-person variability of movement parameters in the WS and TD groups (TD data from Cowie *et al*, 2010). Group means and standard errors shown. LL: leg length.
**Figure 5:** Correlations of variables with scaling performance, measured as riser ht *vs* toedrop slope.

**Table 1:** Additional tests completed by WS participants.
**Table 2:** Group x riser height ANOVAs on movement parameter means in the WS and TD groups (TD data from Cowie *et al*, 2010).
**Table 3:** Group x riser height ANOVAs on movement parameter within-person variability in the WS and TD groups (TD data from Cowie *et al*, 2010).

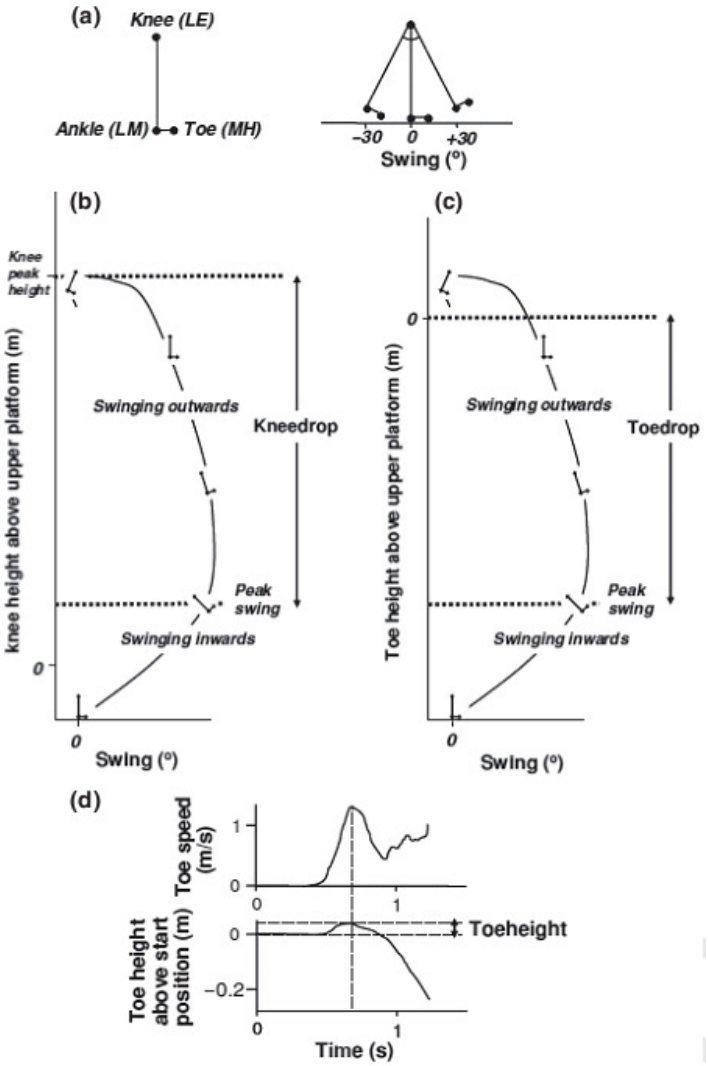


**Figure 1**

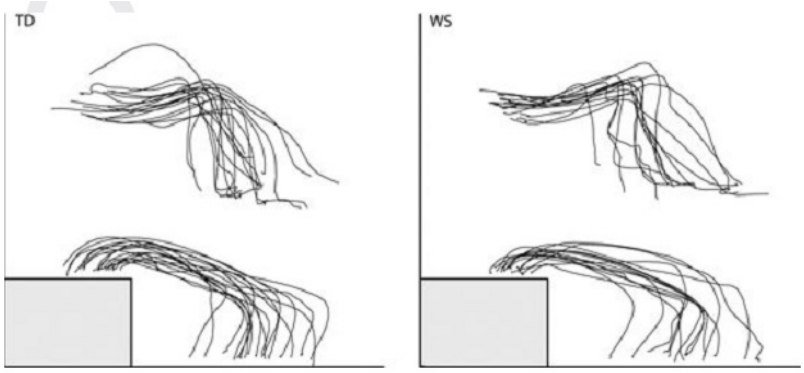


**Figure 2**

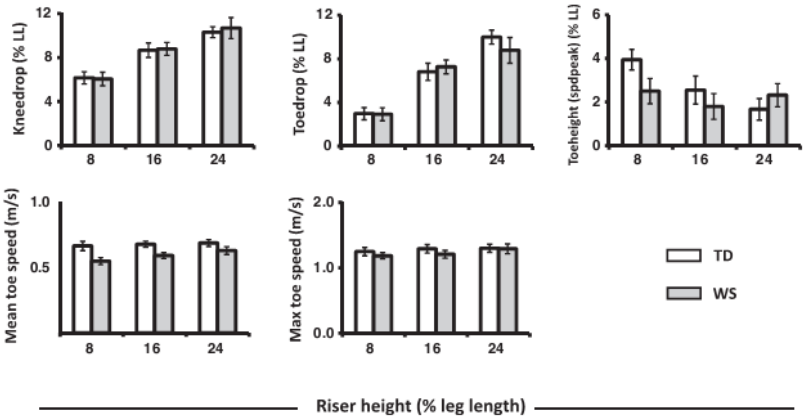


**Figure 3**

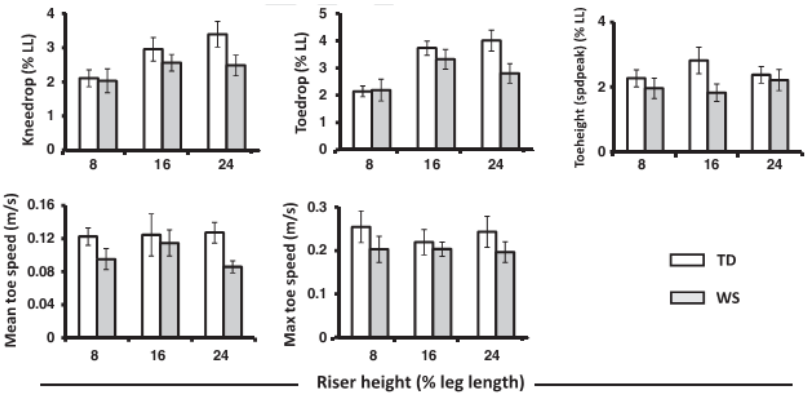


**Figure 4**

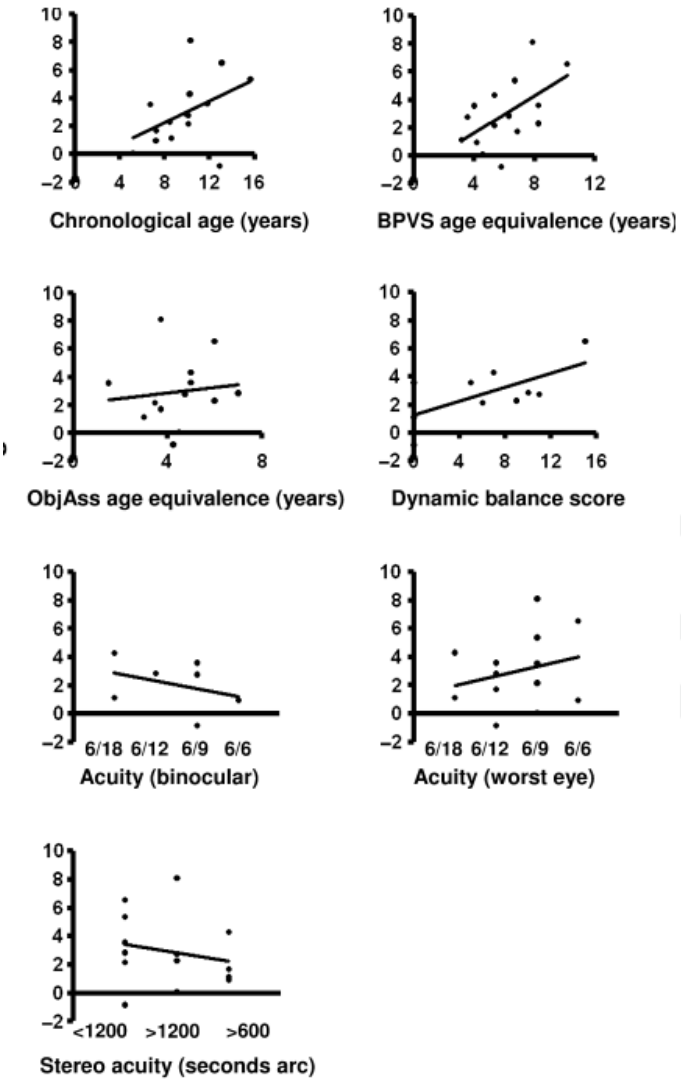
10
8
6
4
2
0
−2
0 4 8 12 16
Chronological age (years)
0 4 8 12
BPVS age equivalence (years)
0 4 8
ObjAss age equivalence (years)
0 4 8 12 16
Dynamic balance score
6/18 6/12 6/9 6/6
Acuity (binocular)
6/18 6/12 6/9 6/6
Acuity (worst eye)
<1200 >1200 >600
Stereo acuity (seconds arc)

**Figure 5**

## Table 1

| Test type | Description | Scoring |
|---|---|---|
| Vision<br>Lang Stereo test *(Lang, 1983)* | Test of stereo vision. Match three shapes defined in a random dot sterogram with disparities of 550", 600", 1200" to corresponding shapes on a test card. | 0: no pictures identified<br>1: 1200" picture identified<br>2: 1200" identified, plus either 550", 600" or both |
| Cambridge Crowding Cards *(Atkinson et al., 1988)* | Test of acuity. At 3 m, correctly identify the middle letter in a display of 5. | Binocular acuity<br>Acuity in worst eye (6/6; 6/9; 6/12; 6/18; 6/24) |
| Motor<br>Movement Assessment Battery for Children: Dynamic Balance subtest *(Henderson & Sugden, 1992)* | From age band 4–6 yrs. Walk along a straight line without feet leaving it. | Number of steps taken in unbroken succession |
| Visuospatial<br>Wechsler Preschool and Primary Scale of Intelligence (WPPSI): Object Assembly subtest. *(Wechsler, 1967/2002)* | Test of visuospatial skills. Assemble pictures from component parts. | Scored according to WPPSI instructions |
| Language<br>British Picture Vocabulary Scale: Short Form *(Dunn, Whetton & Pintilie, 1982)* | Language comprehension. Point to the picture (choice of 4) associated with a word you hear. | Scored according to BPVS instructions |

## Table 2

| | | Kneedrop | | Toedrop | | Toeheight (speed-pk) | | Max toe speed | | Mean toe speed | |
|---|---|---|---|---|---|---|---|---|---|---|---|
| | *df* | *F* | *p* | *F* | *p* | *F* | *p* | *F* | *p* | *F* | *p* |
| Group | 1,31 | 0.005 | .947 | 0.28 | .603 | 0.43 | .519 | 0.47 | .498 | 5.99 | .020* |
| Riser ht (linear) | 1,31 | 69.11 | < .001* | 84.00 | < .001* | 23.60 | < .001* | 3.36 | .076 | 8.00 | .008* |
| Riser ht (quad) | 1,31 | 2.87 | .100 | 8.67 | .006* | 1.44 | .239 | 0.03 | .865 | .001 | .973 |
| Group × Riser ht (linear) | 1,31 | 0.30 | .590 | 0.60 | .443 | 9.25 | .005* | 0.31 | .585 | 2.38 | .133 |
| Group × Riser ht (quad) | 1,31 | 0.005 | .943 | 3.74 | .062 | 0.13 | .721 | 0.78 | .384 | .001 | .973 |

## Table3

| | | Kneedrop | | Toedrop | | Toeheight (speedpk) | | Max toe speed | | Mean toe speed | |
|---|---|---|---|---|---|---|---|---|---|---|---|
| | *df* | *F* | *p* | *F* | *p* | *F* | *p* | *F* | *p* | *F* | *p* |
| Group | 1,31 | 2.23 | .146 | 2.55 | .120 | 3.32 | .078 | 3.26 | .081 | 4.81 | .036* |
| Riser ht (linear) | 1,31 | 8.13 | .008* | 15.13 | < .001* | 0.50 | .483 | 0.10 | .751 | 0.02 | .890 |
| Riser ht (quad) | 1,31 | 1.25 | .273 | 11.79 | .002* | 0.15 | .703 | 0.44 | .511 | 0.70 | .409 |
| Group × Riser ht (linear) | 1,31 | 1.87 | .181 | 3.89 | .057 | 0.09 | .769 | 0.02 | .900 | 0.62 | .438 |
| Group × Riser ht (quad) | 1,31 | 0.05 | .829 | 0.14 | .709 | 1.56 | .221 | 0.71 | .405 | 0.57 | .458 |